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ORIGINAL ARTICLES

SPINA BIFIDA OCCULTA:*

(1) WITH EXTERNAL SIGNS, WITH SYMPTOMS.
(2) WITH EXTERNAL SIGNS, WITHOUT SYMPTOMS.
(3) WITHOUT EXTERNAL SIGNS, WITH SYMPTOMS.
(4) WITHOUT EXTERNAL SIGNS, WITHOUT SYMPTOMS.

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By spina bifida one ordinarily means, and the dictionaries so define it, a cleft or deficiency in the bony column with a lesion of the spinal cord or its membranes—usually a protrusion (meningocele, myelocele, or meningomyelocele). This is, I think, a wrong use of the words. Spina bifida means bifid spine, and that individual has a spina bifida who has a cleft in the spine, whether or not there is protrusion of the spinal cord structures. In 1875, when Virchow[1] reported what was supposed to be the first case observed, he coined the term "spina bifida occulta" to indicate a type of spina bifida in which the lesion was concealed beneath the skin. In a search of the literature as far back as 1825 I found, however, two earlier cases[2] that belonged to this type of spina bifida,† and up to 1910 I found recorded about 85 cases. In these the lesion was indicated

* Read, by invitation, before the New Hampshire Medical Society, May 16, 1917.

† The satyrs of the ancients may well have been suggested by instances of spina bifida occulta with short sacral "tails" and club-feet. Concerning satyrs, see Pauly, *Realencyklopädie der Klassichen Alterthumswissenschaft.*

externally most often by a distinct hypertrichosis over the cleft, somewhat less often by a congenital lipoma symmetrically situated over the cleft, occasionally by a nevus, telangiectasis, or scar, or sometimes by a combination of these. Such a spina bifida occulta is most often found in the lumbar or lumbosacral region, but it may occur in the dorsal or even the cervical region. It usually involves only a few vertebral arches, but it may extend through the greater part of the spinal column. The "woman with the horse's mane,"[3] exhibited many years ago in the "dime museums," with a luxuriant mane of hair over the midline of her back, was an instance of such an extensive bifidity. Mere hairiness of the lumbosacral region, which is noted in many individuals, is not indicative of a spina bifida. Such a well-marked hypertrichosis with long hairs as is shown in Fig. 19, is, however, a pretty safe indication that a spina bifida occulta is present, and probably most of the cases of sacral hair growth studied by anthropologists some years ago as an atavism, or a stigma of degeneracy, are instances of this congenital deformity, as first suggested by Recklinghausen.[4] I have found some very interesting records of such lumbosacral hair growth in several members of a family, however, and spina bifida occulta is not an hereditary phenomenon. The hypertrichosis may not appear, or develop prominence, until puberty.

The congenital lipoma associated with spina bifida occulta is not freely movable, but is somewhat attached to the underlying aponeurosis, and is usually quite circular in outline (Fig. 2). In 4 cases operated upon I have confirmed the interesting observation recorded by A. Jacobi,[5] many years ago, that congenital lipomata differ from the other varieties in being unencapsulated and very finely lobulated, in which respect they are much like subcutaneous fat. Not every case of congenital supraspinal lipoma has a bifidity associated with it.

In several cases of spina bifida occulta scoliosis or other spinal deformity is also present, and sometimes there are other congenital malformations.

In his recent work on the *Surgical Diseases of the Spinal Cord* Elsberg explains the phenomena of spina bifida occulta in the following manner: At birth the cord extends down to the lower end of the spinal column; as the individual grows the cord recedes, relatively speaking, until its lowest end is at about the first or second lumbar arch; if, therefore, any nerve roots are congenitally herniated through a spinal column dehiscence, these will undergo stretching as the disproportion in length between the bony and medullary columns develops with the growth of the individual. Of course, however, this must not be accepted as the entire explanation for the following reasons:

1. In some cases with symptoms there is no hernia of the spinal cord structures.

2. In a few recorded cases, symptoms developed as early as the second year after birth.

3. In some cases, as in one here recorded (Helen H.), the lesion is a teratoma (fibromyolipoma) of the cauda equina.

4. In some cases there is an exostosis in the canal compromising the cord tissues.

5. In many cases there is ample clinical evidence of a lesion in the cord itself.

6. At autopsy in some of the recorded cases dilatation of the spinal canal or degeneration of various tracts has been demonstrated.

At operations and autopsies these abnormalities have been found: A cleft of varying length or breadth in one or more arches accompanied by one of the following conditions: (1) The cleft may give passage to a distinct meningocele; (2) the cleft is closed by a tough membrane adherent to the overlying skin or non-encapsulated fat and connective tissue; (3) the membrane is perforated by a dense band attached to the subcutaneous tissues without and compressing the cord structures within; (4) lipomatous tissue within the canal is concealed by this membrane; (5) the cleft discloses the bulging dura mater; (6) an exostosis within the canal compromises the cord tissues; (7) a myofibrolipoma extends through the cleft into the bony canal distorting and compressing the cord and its nerve roots; (8) dilatation of the medullary canal; (9) degeneration of cord tracts.

The symptoms arising from spina bifida occulta usually appear during adolescence or early adult life, but not rarely they develop during childhood, and occasionally they first appear during middle life. These symptoms may be one or more of the following: Incontinence of bladder or rectum; sensory paralyses; motor paralyses; disturbances of the reflexes; trophic ulcerations and gangrene. Symptomatically, therefore, spina bifida and spina bifida occulta are one. It must be noted, too, that pathologically spina bifida and spina bifida occulta do not greatly differ. Tumor tissue sometimes accompanies evident spina bifida; hypertrichosis sometimes does; an evident meningocele is occasionally partially concealed by a congenital lipoma; and, finally, there are reasons for believing that a spina bifida sometimes shrinks, leaving only scar tissue or other index to a spina bifida occulta.

Of the type of spina bifida occulta with symptoms and with external signs, by which I mean lipoma, hypertrichosis or nevus, the following 3 cases are interesting examples.

GROUP I.—*Spina Bifida Occulta, with External Signs, with Symptoms.*

Sigrid H., female, aged eighteen years, came under my care in December, 1907, for an extensive ulceration of the inner and flexor aspects of the great toe of the left foot, indolent in appearance, and with foul discharge.

She was a native of Jönköping, Sweden, where, at the age of seven, the second left toe was ulcerated from what was presumed to be frost-bite. This ulceration recurred each winter and, at the age of twelve, partial amputation of the toe was required.

Three years later (aged fifteen) the stump of the toe ulcerated and the phalanges necrosed. Six months later the fifth toe ulcerated, refused to heal, and was amputated.

She never had vesical, intestinal, or motor disturbances. She never noticed anesthesia in either foot or leg. No lightning pains, but these ulcerations were mildly painful. At the appearance of each fresh ulceration she said she had malaise, headache, nausea, and fever, and the toe would first appear red.

Observations of this patient and the next one to be reported confirmed this statement: preceding and accompanying each fresh ulceration, both patients felt ill and had a rise of temperature to 103° or 104°. The appearance of a fresh ulceration could thus be accurately predicted.

The girl was referred to my surgical from the neurological department of the Mount Sinai Hospital Dispensary as a purely surgical case, and with the assurance that there was no neurological lesion. I mention this not in criticism, but to illustrate how even a trained neurologist may overlook a spina bifida occulta until a single experience teaches him to be on the lookout for it.

The foul ulceration for which the patient came to me has been briefly described. The first phalanx was necrotic. The wound was laid open and this bone was removed. Slowly the wound healed, *without granulating*, after the head of the first metatarsal spontaneously discharged. Specific medication seemed to exercise no influence. The dorsalis pedis, popliteal and posterior tibial vessels pulsated normally. The urine was negative. The foot condition and the patient's nativity suggested the possibility of leprosy.

Dr. Prince A. Morrow, who was kind enough to examine the patient for me, wrote: "I think the diagnosis lies between syringomyelia and leprosy, probably the latter. I have seen quite a number of unilateral manifestations due to leprosy, although the dystrophic changes are usually symmetrical. The rather characteristic plantar ulcers and the anesthetic ulcers are in favor of leprosy."

In April, 1908, the two remaining toes (third and fourth) ulcerated and the two last phalanges of these were removed, leaving stumps. The foot was then quite edematous but not painful, although there developed at this time a plantar ulcer near the base of the third toe (Fig. 1). Smears from the ulcers showed no lepra bacilli.

I had the patient admitted to the second surgical division of the Hospital (Dr. Lilienthal) for further study. I abstract from her history the following positive findings:

Over the lower lumbar and upper sacral region extending down

from the third lumbar vertebra is a circular tumor about four and a half inches in diameter symmetrically situated over the midline

FIG. 1.—Sigrid H., September, 1908. Plantar ulcers at base of third toe and over the head of the second metatarsal. Note stumps of toes.

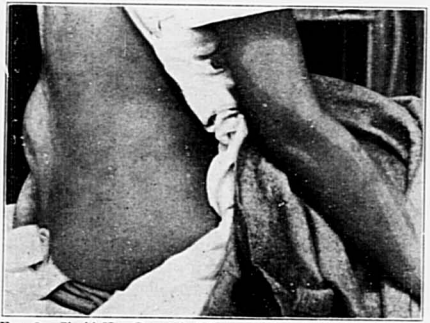

FIG. 2.—Sigrid H. Congenital lipoma over spina bifida occulta.

(Fig. 2). It is about one and a half inches thick in its center, of doughy consistency, painless and not tender, moderately movable over the deep structures and adherent to the skin at its center. This

tumor was present since birth and has grown with the patient. In the spine no cleft can be felt. No hypertrichosis. The left foot is as above illustrated and described. Blackish, rugous epithelium covers part of the third toe where it abuts against the great toe. The plantar ulcer is small, deep, conical, lined with granulations, but does not extend to the bone. Reflexes: abdominal—right, active; left, absent: gluteal—absent on both sides: knee-jerks—right, feeble; left, absent: achilles—right, feeble; left, absent. No Babinski or

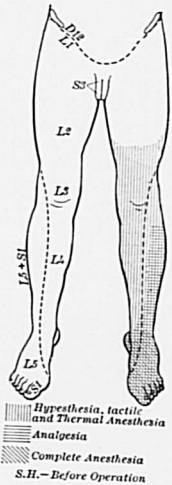

FIG. 3

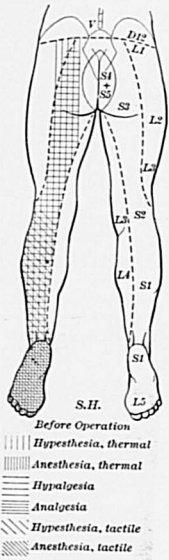

FIG. 4

Oppenheim phenomenon. Motor power of lower extremities normal and equal. Gait normal. No ataxia. Tests show sensory paralyses of the left lower extremity as illustrated in Figs. 3 and 4. Roentgenogram shows a cleft in the *left* lamina of the fourth lumbar arch. The fifth lumbar arch is not of normal shape (Fig. 5). Clinical diagnosis: spina bifida occulta associated with congenital lipoma over the lower end of the spine; cleft in the fourth lumbar arch with a lesion involving the posterior fourth and fifth lumbar and first, second, and third sacral roots or cord segments.

Fig. 6, taken from Sobotta's *Anatomy*, illustrates how pressure on the cauda equina at the level of the fourth lumbar vertebral arch would affect the posterior roots below the level of the third

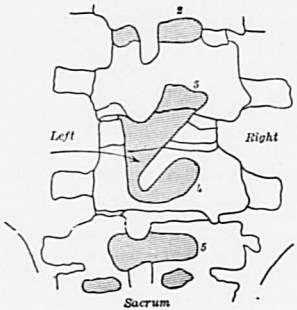

Fig. 5.—Sigrid H. Tracing of roentgenogram of lateral spina bifida.

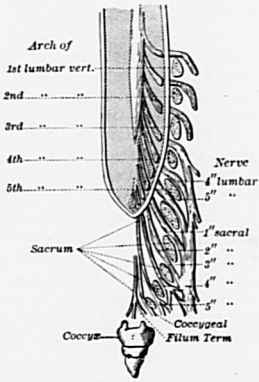

Fig. 6.—Emergence of the posterior nerve roots between the bony arches. Illustrating that pressure on the cauda equina at the level of the L4 arch would affect these roots below the L3 root. (After Sobotta.)

lumbar root, and how, if sufficiently circumscribed, some of the lumbar and sacral roots on one side only might be involved. This was, in fact, just the condition found at

Operation (Brickner and Elsberg), July 3, 1908. Curvilinear incision over the upper border of the tumor, which was found to be a large collection of unencapsulated fat arranged in very small lobules like subcutaneous fat and practically free from bands of fibrous tissue. Incision was carried through this down to the lumbar aponeurosis and the spinous processes. These were found normally disposed, but just to the left of the fourth lumbar spine there was a distinct hiatus in the muscles—an opening lined with fascia which here dipped down toward the spinal canal. This opening easily admitted the finger-tip. Through it issued a strong, apparently dense columnar structure. This structure terminated in the lipoma, but it had no attachment to the skin. With some of the attached fat it was dissected free and dragged through the opening. Incision through the fibrous envelope revealed a serous lining, within which were several nerve fibers. A probe could be passed along these into the spinal canal, and cerebrospinal fluid escaped alongside it. The dural extension thus exposed was further opened, revealing that the hernia was a meningeal sac containing four or five distinct nerve roots of the cauda equina, which bent upon themselves and reëntered the spinal canal. At the point at which these roots were flexed they were found somewhat adherent to the sac, which was at this point much constricted. Distal to this point the meningocele then expanded, but no fluid was noted in this expansion.

The sac was removed, the nerve roots were reduced into the spinal canal, and the dura was sutured over them. The lumbar aponeurosis was closed over the hiatus by a plastic operation.

Following the operation the patient complained of numbness in both lower extremities for a few days, but there was no other untoward effect, and the wound healed *per primam*. Soon after the operation the patient volunteered the statement that her left lower extremity felt more normal, more like the right, than it had for many years before, and improvement was noted in the sensory paralyses, as indicated in Figs. 7 and 8.

Pathological examination of the amputated sac and of the adherent fat revealed no features of special note.

The patient was discharged from the hospital August 4 and readmitted August 25. The ulceration in the third toe had progressed and the plantar ulcer had also increased in size, exposing the head of the second metatarsal bone.

By September 28 these ulcers were healed without any bone necrosis, and the patient was again discharged with a raised sole on her right shoe and crutches.

In November the plantar ulcer reopened, as usual with a rise of temperature to 103°. Surrounding the ulcer there developed a thick, soft callus in spite of the fact that the patient had not put her foot on the ground for many months. During the next few

months the ulcers improved and relapsed and others developed. The patient was readmitted to the hospital.

July 26. Typical Pirogoff amputation (Brickner). All the bloodvessels appeared quite normal. Primary union.

Pathological report of the amputated foot showed—to summarize—practically normal vessels, but marked bone atrophy. This atrophy was found in portions of the bone remote from the inflammatory process. The nerves of the foot showed no degenerative changes. In the immediate vicinity of the ulcers there were moderate inflammatory changes in the nerves.

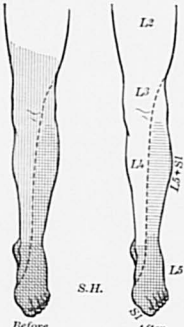

FIG. 7

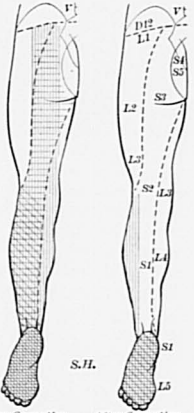

FIG. 8

Contrary to advice the patient insisted upon wearing a shoe containing a cork foot. The pressure of this cork against the anterior aspect of the stump caused an ulceration of the skin within a month after the patient was discharged from the hospital.

Between October, 1909, and May, 1910, the patient neglected treatment, reporting but rarely, the skin necrosis meanwhile extending into a large shallow ulcer surrounded by a dense callus. The raw area was granulating, bled easily, and would probably have healed under good treatment if the patient would have abandoned the shoe, which she insisted upon wearing.

I lost track of her between May, 1910, and January 30, 1911, when she reported again. The ulceration was then much reduced in size, but a gangrene of the heel portion, extending almost to the bone, was present.

February 6, 1911. Osteoplastic amputation through the leg (Brickner). Primary union. Discharged from the hospital.

Soon after that I lost all trace of the patient until in the winter of 1917, through the courtesy of Dr. John Moorhead, I saw her again in the Harlem Hospital. Since my last preceding note she had married and had two healthy children, free from deformity. She had no difficulty in parturition, had no bladder or rectal trouble. During the past year she had lost the fourth and fifth toes of the *right* foot following infection of a corn. She had gone to the Harlem Hospital because of a deep indolent ulceration of the amputation stump (left leg), which had been increasing in size for the preceding six months. During all these six years she had worn an artificial leg pressing against the stump and from time to time had had superficial ulcers, which, however, healed. This deep ulceration did not extend to the bone, and an *x*-ray picture showed that the bone was not at fault.

Sensory examination in March, 1917, showed in the outer half of the stump below the knee, thermal and tactile hypesthesia as previously, but the right foot and the lower portion of the right leg posteriorly now also show thermal and tactile hypesthesia. The large, deep ulcer of the stump gradually healed.

Whether or not the release of the nerve roots in the spinal hernia obviated any other troubles (*e. g.*, vesical) it would be mere speculation to say.

Our experience with this case suggested to Dr. Elsberg the diagnosis in the case of the following patient, whom he had had under observation for several years at Montefiore and Mount Sinai Hospitals:

Helen H., aged twenty-six years, single, hairdresser. No member of the family, direct or collateral, had had a similar trouble. One sister has a congenital club-foot, unilateral; no other congenital deformities in the family.

Patient entirely well until nineteen. Then, in 1901, she noticed two suppurating ulcers, the size of dimes, in the calf of the left leg, which had appeared spontaneously and painlessly and healed painlessly in two weeks. A month later severe shooting pains in the calf of the left leg and especially the left foot. These continued for a few weeks, when ulcers appeared on the fifth toe and on the sole of the left foot, which swelled. These healed and broke open again repeatedly during the next nine months, and the toe was then amputated. Sensory paralysis was then noted in the other toes, and later in the left foot, leg, and thigh. During the next few years she similarly lost the second, third, and fourth toes, and had ulcers

of the sole and heel of the same foot. She had no urinary disturbances, but had had diarrhea for several weeks before admission to the hospital, July 20, 1908.

Physical examination revealed the following positive findings: Spine normal to inspection and normally flexible. Over the upper end of the sacrum and the fifth lumbar vertebra is an almost inconspicuous swelling, due apparently to a small accumulation of fat; it has no distinct boundaries, but extends almost to the sacro-iliac joints. It is asymmetrical in depth, being thickest on the right side. The back is covered with light colored lanugo, but over the left side of this lipoma there is a small tuft of very small, short, black hairs, about a dozen in number, about 1 cm. in length, and very slender. The lumbar spinous processes are easily palpable and appear well formed except the fifth, which feels small and lies a trifle to the right of the line through the others. Just below the fifth lumbar process, viz., at the site of the above-mentioned swelling, the fingers on deep pressure sink into a shallow depression, and bony contact is not transmitted. The rest of the sacrum and the coccyx appear normal to palpation.

FIG. 9.—Helen H. Tracing of roentgenogram of spina bifida.

There is a sacrococcygeal dimple, but no sinus. Roentgenogram shows the spina bifida illustrated in Fig. 9. It consists of a distinct and extensive hiatus in the first sacral arch, most marked on the left side. The fifth lumbar arch is complete, but it is asymmetrical and narrow, the spine lying to the right of the midline.

The lower extremities show an atrophy of the left calf and exaggeration of the left knee-jerk. No ankle-clonus on either side. The outer side of the single remaining toe of the left foot is practically

denuded of epithelium from a recent bleb. There is a plantar callus over the head of the second metatarsal. Scars of ulcers on the calf and over the tendo Achillis. Locomotion and station normal. Abdominal reflexes lively and equal. The right lower extremity shows no disturbances of sensation. The left lower extremity and left side of the anogenital region show the sensory paralyses illustrated in Figs. 10 and 11, indicating, with the trophic disturbances, an involvement of the fifth lumbar and all the sacral nerve roots or segments on that side and a slight involvement of the second, third, and fourth lumbar roots.

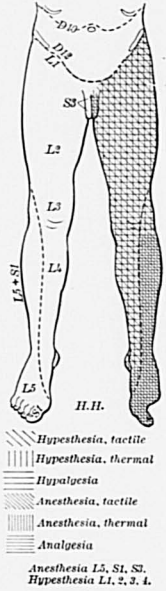

FIG. 10

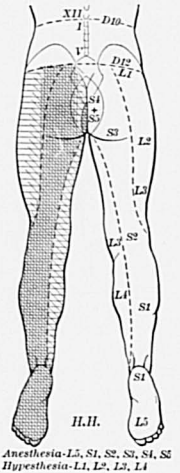

FIG. 11

Operation (Elsberg and Brickner), July 27, 1908. Slightly oblique transverse incision across the upper level of the sacrum carried down through the underlying fat, unencapsulated and finely lobulated, to the lumbar aponeurosis. Over the upper end of the sacrum this aponeurosis was found to dip in somewhat like a shallow funnel, but

there was no opening in it and no structure passed through it. The depressed portion of the aponeurosis was circumcised, and this was dissected up from the underlying muscle, which was retracted, exposing a mass of fat and connective tissue extending down between the muscles in the midline and firmly attached to the under surface of the lumbar aponeurosis. No dura or congenital band was here encountered, but further dissection showed that the mass of fat and connective tissue stretched down to the pulsating dura mater seen through the large defect in the first sacral arch. This mass was dissected from the dura at the point where it found attachment. The dura was found to be not adherent to the overlying laminæ in the neighborhood of the hiatus. The dura was then incised, cerebrospinal fluid escaping. Within the dural opening several nerve roots were seen floating in the fluid, some lying against the inner surface of the dura, but it was demonstrated that they were not attached to it. The dura was sutured and the hiatus was closed by a plastic operation.

Microscopic examination of the removed mass of fat, connective tissue, and aponeurosis showed no other structures within it.

The patient vomited and had headaches for several days after the operation, and required catheterization. Primary union. Involuntary urination developed for a few days and transitory diarrhea recurred.

Sensory examination a few weeks after the operation showed some improvement in the left lower extremity, as indicated in Figs. 12 and 13. She was discharged August 22, 1908. During the next few months she complained much of pain in the back. The pains in the left leg had, however, gradually diminished.

In October, 1908, there developed a plantar ulcer surrounded by callus, which healed very slowly. During the next few months she complained much of severe pains in the left groin and the back. During the following year recurrence of ulcers in the sole of the foot and calf, always preceded and accompanied by fever.

Operation (Elsberg and Brickner), May 16, 1910. Lumbar laminectomy—first, second, and third lumbar laminæ removed. Dura opened and the exposed cord revealed the following: Its arteries were more prominent than normal. Much of the pia was lifted from the cord by coagulated cerebrospinal fluid. The cord proper extended farther down than usual, namely, to the third lumbar arch. The coagulated fluid was liberated, but no further lesion was noted in the cord or its roots, anterior or posterior, at this level. Laminectomy then continued down to the spina bifida, removing the fourth and fifth lumbar arches. There was then revealed at about the level of the fifth lumbar arch what appeared to be a lipoma of the cauda equina, occupying all of the spinal canal at this situation. It was soft, yellow, and of irregular conformation. Small pieces were removed for histological study, but it was impos-

sible to dissect the fat from the roots of the cauda equina, which ran through it and were intimately attached to it. The posterior fifth lumbar nerve root on the left side was then divided to relieve pain. Histological examination of this fatty tissue showed that it was a fibromyolipoma and contained a number of fine nerve bundles, some of which were apparently ramifications of larger nerves lying in the connective tissue. Most of the nerve bundles showed proliferation of the endo- and perineurium, and some of those lying in the fat were almost completely converted into fibrous cords. The connective tissue was rich in capillaries and arterioles, in most of which there were leukocytes, with perivascular aggregation of white cells. Some of the scattered striated muscle fibers showed degeneration. (Examination by Dr. I. Strauss.)

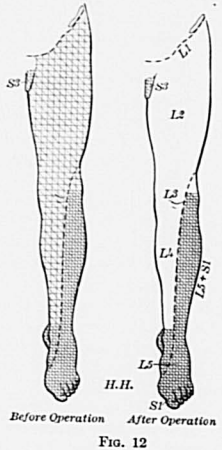

FIG. 12

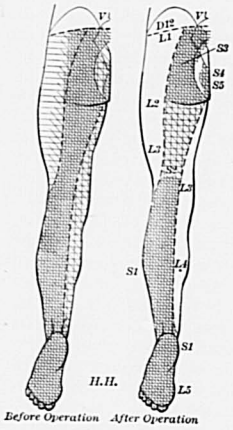

FIG. 13

Since this operation the patient's symptoms have progressed. A few months after it she developed drop-foot on the left side and subsequently also on the right side. She has very poor control of the bladder or rectum, complains much of backache, has cystitis from time to time, has frequent attacks of mucous colitis and frequent recurrences of the ulcerations. She walks but little, and out of the house is obliged to use a wheel chair.

These two cases are especially interesting in the laterality of the bifidity and, for a long period of years, of the symptoms. *They are interesting also in developing the observations concerning the*

growth of callus as a purely trophic phenomenon independent of pressure, and the prostration and marked rise of temperature which preceded and accompanied the outbreak of an ulceration, as we repeatedly noted.

Sam. S., male, aged thirty years, suspender-maker, was admitted to the dermatological service at Mount Sinai Hospital July 15, 1915, with an ulceration, about two inches by three inches in extent, on each buttock, symmetrically situated two and a half inches from

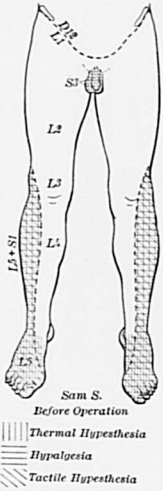

FIG. 14

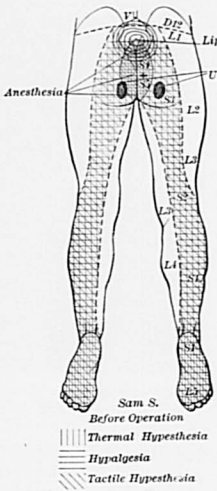

FIG. 15

the anus. These punched-out, freely discharging ulcerations had healed and recurred three or more times in a period of two years. The patient also had a purulent cystitis, of unknown origin, a right inguinal hernia, a peculiar falsetto voice, and a lipoma over the lumbosacral area characteristic of spina bifida occulta. He had no motor or rectal symptoms, but there was symmetrical sensory paralysis of the anoscrotal regions, buttocks, and lower extremities, as shown in Figs. 14 and 15. No alteration of the reflexes. Wassermann reaction negative.

A roentgenogram showed a narrow cleft in the fifth lumbar arch and an absence of all the sacral arches (Fig. 16).

Under local treatment in bed the ulcerations healed and the patient was discharged, under observation, August 27, 1915.

When he reported again, ten days later, the ulcerations had recurred as before. He was then referred to me for operation and admitted to Dr. Elsberg's service.

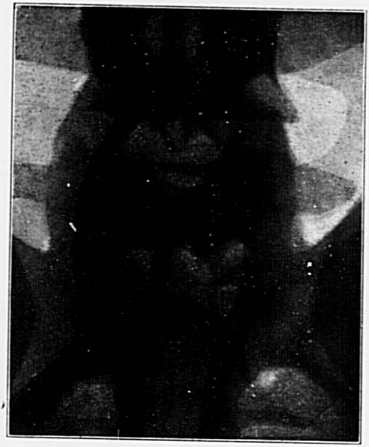

FIG. 16.—Sam S. Roentgenogram showing cleft in L5 arch and wide hiatus in all the sacral arches.

Operation (Brickner), September 10, 1915. Semicircular incision over the upper and left border of the mass, a baseball-sized flattened lipoma, characteristically congenital in its fine lobulation and absence of capsule. It was cautiously dissected out. The wide sacral dehiscence was found covered throughout with a dense aponeurosis except at its upper end, where a small sac protruded into the surrounding fat, to which it was adherent. This sac was found to contain a few nerve roots so adherent to its inner surface that they could not safely be dissected from it. The herniated mass was a little larger than a thimble and escaped through a finger-tip-sized opening in the spinal canal. Below this opening the aponeurosis was incised to permit exploration. Only fat was seen, extra- and intradurally. Some of this was removed. The protruding nerve roots and attached dura were reduced into the canal *en bloc* and the opening was closed over firmly by a plastic operation on the aponeurosis.

Nine days later the wound was healed *per primam*, the ulcers were almost healed, and there was a decided improvement in the sensory disturbances. This improvement has continued (Figs. 17 and 18), and today practically nothing remains of the sensory paralysis—formerly so marked in the anogenital and gluteal regions—except thermal hypesthesia of the feet. I operated upon this patient a year ago for his inguinal hernia, and I have kept him under observation since the spinal operation two years ago, during which time the buttocks have remained solidly healed. Six months ago he had an infected abrasion of the dorsum of the right foot, but it healed normally.

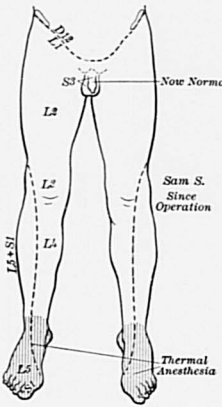

FIG. 17

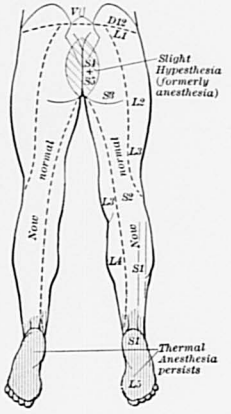

FIG. 18

Even if the continued cure of the previously recurring ulcers is a mere coincidence—which I cannot admit—the pronounced change in the sensory phenomena bespeaks a real benefit from his spinal operation.

GROUP II. *Spina Bifida Occulta, with External Signs, without Symptoms.* It would seem probable, especially from a study of the literature of abnormal hair growths, that some individuals with spina bifida occulta, even though marked by a very decided hypertrichosis or a congenital lipoma, go through life without any symptoms, or, at any rate, without any symptoms that bring them to the physician's attention. The following is such a case, although the patient died before he was twenty-three years of age.

Harold H., male, aged twenty-one years, was referred to me by Dr. Bullowa, and was in Dr. Elsberg's service at Mount Sinai Hospital in 1914 with a chondrosarcoma of the head of the tibia. Resection of the knee (Brickner), recurrence, amputation of thigh (Brickner). Intrathoracic metastasis. Died at Roosevelt Hospital, November 18, 1915. No motor, trophic, or sensory disturbances were noted, but he had a localized, bushy growth of long, straight hairs over the lumbosacral region (Fig. 19), and a roentgenogram showed a wide cleft in the fifth lumbar arch and a wide hiatus in the first, second, third, and fourth sacral arches (Fig. 20).

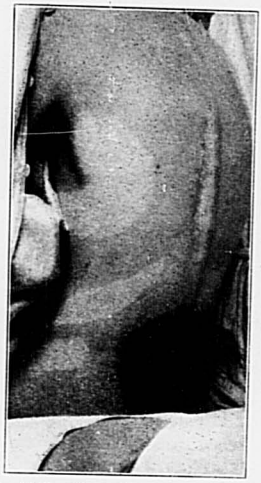

FIG. 19.—Harold H. Hypertrichosis over spina bifida occulta.

R. S., female infant, aged three months, admitted to Dr. Moschcowitz's service at Mount Sinai Hospital August 18, 1916, with a congenital lipoma over the lumbar spine, small at birth but gradually increasing in size to that of a large crab-apple. No defect in the vertebral arches could be felt through it, and pressure upon it caused no symptoms. Physical examination otherwise negative.

August 22. Operation for spina bifida occulta lumbosacralis (Brickner). The swelling was partly circumcised and the lipoma, finely lobulated and non-encapsulated, was cautiously dissected up. A cleft was palpated in two arches, probably L5 and S1, closed over

by a tough membrane (membrana reuniens) except at one spot, through which fat extending from within the vertebral canal became continuous with the extravertebral lipoma. The membrane was incised to permit exploration, exposing much white fat lying upon and among the posterior roots of the cauda equina, which appeared normal. Much of this fat was picked out and the herniated portion

FIG. 20.—Harold H. Narrow cleft in L5 arch and wide hiatus in (almost complete absence of) S1 arch.

was removed. The membrane was closed with sutures, and a plastic closure of the lumbar aponeurosis was made over this. The baby was discharged, well, September 15.

Although the infant had had no symptoms the operation was done as a means of preventing, if possible, the development of symptoms later in life.

GROUP III. *Spina Bifida Occulta, without External Signs, with Symptoms.* This is the group to which I especially call attention, for it has hitherto scarcely been described.

In April, 1909, when I demonstrated before the surgical section of the New York Academy of Medicine the first and second cases here reported, the almost insignificant lipoma in the second case led me to conclude by urging "a careful scrutiny of the back and an *x*-ray picture in all cases in which there are incontinence of urine or feces or, in the lower extremities motor, sensory, or trophic disturbances, the cause of which is not otherwise evident."[6] I find that in the same year, 1909, Alfred Fuchs[7] published a study of 24 cases of enuresis in adolescents and adults, in which he described the following conditions, variously associated, as due to a congenital lower cord lesion which he called "myelodysplasia:" (1) Sphincter weakness, especially the so-called neurosis, enuresis nocturna. (2) Well-developed syndactyly between the second and third toes, and sometimes the fourth toe, in 66 per cent. of the cases. (3) Dissociated sensory disturbances in the feet, especially the toes, but also at the outer side of the feet, and chiefly thermal hypesthesia or anesthesia, in 75 per cent. (4) Defect in the sacral canal varying from an abnormally high opening of the hiatus sacralis to a spina bifida. Fuchs found these vertebral anomalies roentgenographically in 6 out of 10 cases of enuresis, and regards them as a rudimentary spina bifida occulta development. (5) Disturbances of skin and tendon reflexes, 11 of the 24 cases. (6) Foot deformities—percentage not stated.

Georg Peritz[8] agrees with Fuchs that the association of enuresis and spina bifida occulta sacralis is not merely accidental, thus disagreeing with Tromner and also with Lewandowsky, who, in his *Handbuch der Neurologie,* says that enuresis and spina bifida occulta are on the same degenerative basis, but have nothing to do with each other. In 22 adolescent cases of enuresis Peritz found spina bifida occulta roentgenographically in 68.2 per cent. In 20 children with enuresis Peritz found spina bifida occulta roentgenographically in 35 per cent.

William G. Spiller[9] reported a case of enuresis increasing, and motor weakness and sensory disturbances developing, after moderate exercise, in the lower extremities, in which the roentgenogram showed a grave defect of the sacrum and lower lumbar vertebræ, the back appearing entirely normal to sight and palpation.

Palmer Findley[10] renews attention to the fact that "virginal prolapse of the uterus occurs with greatest frequency in the newborn, and in most instances there are associated congenital deformities, notably spina bifida," and indicates that it would appear to be also a contributing factor sometimes in prolapse of the uterus in multiparous women.

In 17 cases of congenital prolapse of the uterus Ebeler and

Duncker[11] record that 15 had spina bifida; and they found occult spina bifida roentgenographically in 23 out of 28 multiparous women with prolapsus uteri. They also found it in 3 of 28 multiparous women without prolapsus.

The following 3 cases may be cited to illustrate this group. Palpation may or may not give sufficient indication of spinal arch deformity or dehiscence. Roentgenography shows it clearly.

Mollie C., unmarried, female, aged twenty-one years, was referred to me by Dr. Leon Lesser in December, 1915, for a partial incontinence of bladder and rectum, which existed since she was nine years old. She talked when one year old but did not walk until four. When in the street she often soils herself with urine or feces; at home she is able to reach the toilet in time to prevent this mishap. Her chief complaint is that she therefore must remain indoors most of the time. She urinates five or six times daily and once or twice during the night. Usually she has no diarrhea.

There were no other symptoms. The urine was normal. Physical examination was entirely negative. I found no sensory, trophic or motor disturbances and no abnormality of the reflexes. Nevertheless it occurred to me as possible that, in spite of the absence of any external sign, there might be an occult spina bifida. (I was not then familiar with the articles of Fuchs and of Peritz.) A radiogram showed a cleft in the third and fourth sacral arches (Fig. 21).

I had the girl admitted to Dr. Elsberg's service at Mount Sinai Hospital, where he and I observed her for a week, but no new data were developed. Cystoscopic examination showed no bladder lesion. In this case the limited character of the symptoms did not appear to urge the desirability of a spinal exploration. There is obviously no hernia of the cauda equina. The lesion is perhaps in the cord itself. Her condition has not improved during the past year and a half, however, and perhaps an exposure of the cauda equina may reveal some roots adherent to the membrane across the cleft, the release of which may be beneficial.

Benjamin T., male, aged fifty-eight years, was admitted to Dr. Moschcowitz's service at Mount Sinai Hospital June 3, 1916.

Twenty-five or more years before he developed a plantar ulcer of the left foot—an infected wound arising, he thought, from a shoenail. The foot became much swollen and the ulceration remained unhealed for several years. The infection gradually involved the first phalanx of the great toe, which was removed. Two weeks later the fourth toe became involved and the first and second phalanges were removed. Over the finally healed plantar sore callus formed, which the patient frequently pared down. For thirteen years the foot remained healed. Then a perforating ulcer reappeared at the same plantar site, and was finally healed. It

recurred four years later and persisted again for a long time before healing was effected, after treatment at the Vanderbilt Clinic and Har Moriah Hospital. Six months later it again recurred and involved the third toe, which was removed by Dr. Moschcowitz. This was followed by healing.

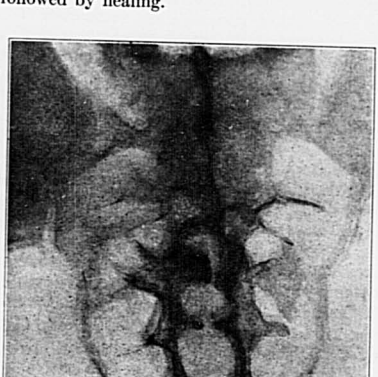

FIG. 21.—Mollie C. Sacral spina bifida occulta. Narrow clefts in the S3 and S4 arches.

Three months before admission to the hospital a painless blister, then an ulcer, appeared on the second toe, increased in size and depth, and extended into the metatarsophalangeal joint. The ulcer was the size of a dime, with sharp borders. The toe and the corresponding metatarsal region were dull red and swollen. There was slight tenderness. In a callous area over the ball of the foot was a linear scar 2 inches long. The fifth toe appeared normal; the first and the fourth had only the terminal phalanx; the third toe was missing; the second has been described.

The appearance was not that of arterial disease. The dorsalis pedis and posterior tibial pulsated normally on both sides. There was no "blushing" or "paling" phenomenon. The urine and heart were normal. Wassermann reaction negative. Blood-pressure 150 to 170. Emphysema. Gait and station normal. No bladder, rectal, or motor symptoms. The neurologist reported "a negative neurological examination. The condition does not depend upon any neurological disturbance."

It seemed to both Dr. Moschcowitz and me, however, that the phenomena were trophic in origin, and I suggested a roentgenographic examination of the lower spine for a bifidity. The sacral spinous processes were not palpable. Indeed, there appeared to be a depression involving the lower half of the sacrum. The roentgenogram showed an extensive sacral hiatus (Fig. 22). There is, in fact, thermal hypesthesia of both feet, although no apparent tactile hypesthesia or hypalgesia.

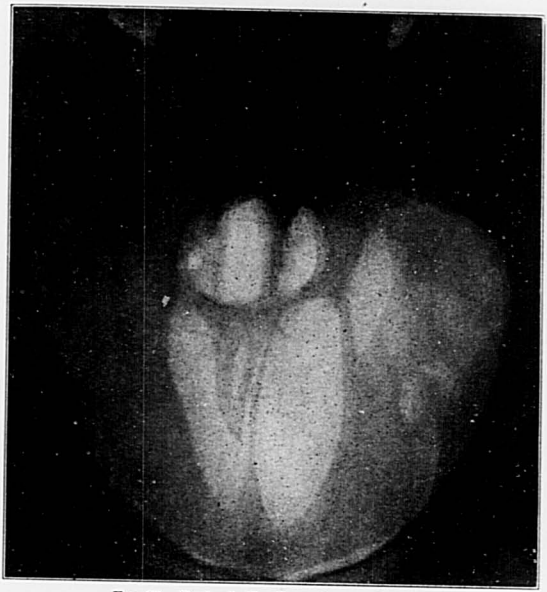

FIG. 22.—Benjamin T. Extensive sacral hiatus.

The third toe was amputated. With the healing of this wound there were no further symptoms and an exploration of the spine was considered unjustified. The patient is, however, under observation.

The following case I present quite tentatively, for the diagnosis is doubtful.

Max H., male, aged fifty-two years, on the neurological service of Dr. Sachs at Mount Sinai Hospital, January 17 to February

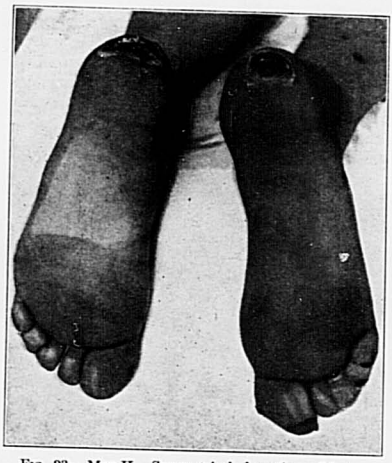

FIG. 23.—Max H. Symmetrical ulcerations of feet.

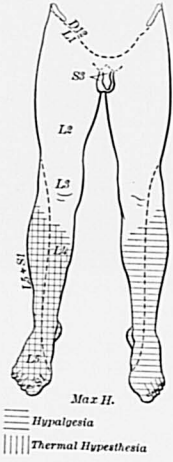

FIG. 24

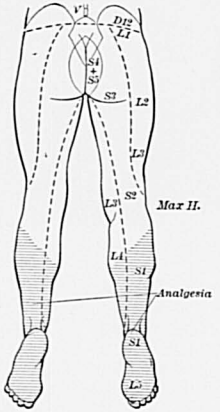

FIG. 25

24, 1917. Ten weeks before admission he had burning in the feet radiating into the legs, sometimes with tingling sensations in the finger-tips. Four weeks later ulcers developed on the feet, painless, and not tender. For some time he had been urinating very frequently, every half-hour during the day and twice at night; no shooting pains or trouble in walking.

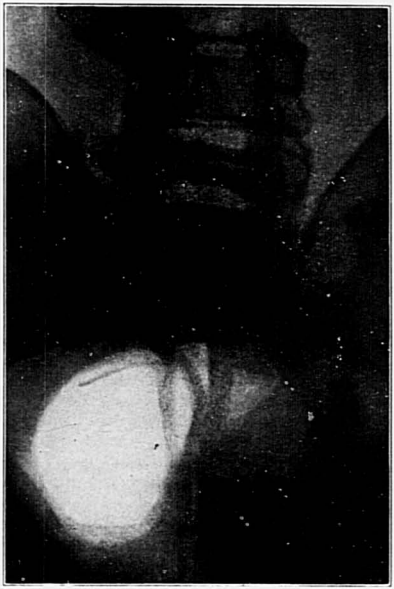

FIG. 26.—Max H. Extensive sacral hiatus.

Examination gave the following positive findings: Hands cyanosed, cold and clammy; coarse tremor. The feet are cold, slightly cyanotic, perspiring. They show the following symmetrical ulcerations (Fig. 23): On each heel an area of dry gangrene about the size of a quarter; on the inner aspect of each big toe a gangrenous area the size of a dime; the right big toe shows also a gangrenous area the size of a nickel. There is ulceration about the nails of the second and third toes of the right foot. Pulsation of the dorsalis pedis artery is readily palpable in both feet. Hypalgesia and

analgesia are noted in both extremities, as shown in Figs. 24 and 25. Cerebrospinal fluid: 11 cells, 32 per cent. lymphocytes, 68 per cent. polynuclears; Wassermann reaction negative. Urine negative. Roentgenogram shows extensive sacral hiatus involving all of the arches except the first (Fig. 26). Electrical reactions normal.

The neurologists inclined to the diagnosis of a neoplasm either just above the conus or in the cauda equina. Dr. Sachs made the following note in the history: "Differential diagnosis between spinal gliosis with possible cavity and a neoplasm of the cauda equina. The onset thirteen weeks ago with sensory phenomena, rapid ulcer formation with gangrene, with preservation of gross muscular power and absence of marked vesical symptoms, argues in favor of neoplasm rather than gliosis. Even if gliosis were the condition, exploratory operation is advisable." Dr. Abrahamson believed that the lesion is a tumor of the type found in spina bifida occulta. Dr. Strauss seemed more inclined to think that the spinal bifidity is an accidental association and that the lesion is a cord tumor just above the conus.

February 17. The patient had a hemoptysis and developed dulness at the right apex. About the same time it was noticed also that there was diminished power in the right lower extremity. He refused operation and left the hospital unrelieved.

None of my 3 cases of spina bifida occulta without external signs with symptoms, and none of the cases of enuresis, etc., recorded by Fuchs, Peritz, Spiller (and one by Alfred Sänger[12]), has been submitted to operation, and a lesion of the cord structures corresponding in type to a spina bifida has therefore not been demonstrated.

Admittedly in some cases at least the bifidity of the spine may be a congenital abnormality that has no etiological relationship with the symptoms, but in many cases, and especially in the case of Benjamin T., with the fairly characteristic progressive ulceration of the toes of one foot and the thermal hypesthesia of both feet, the presumption is very strong that the lesion at fault is indeed a spina bifida occulta.

Group IV. *Spina Bifida Occulta, without External Signs, without Symptoms.* For some years I have been interested in observing that a considerable number of roentgenograms of the lower spine of patients exposed for examination of the urinary tract, the hip, etc., shows a cleft most often in the fifth lumbar arch or in one or more of the sacral arches. In what percentage of apparently normal individuals this anomaly is found roentgenographically I cannot say, but I have the distinct impression that it is not very uncommon. Perhaps a careful inquiry into some of these cases might show some disturbance of bladder control or loss of thermal or tactile sensitiveness in the lower extremities; but in the absence

of such a study I think we may be prepared to believe that there is found in a certain proportion of healthy individuals a congenital defect in the lumbar or sacral arches without any affection of the spinal cord tissues. The following cases are illustrative.

FIG. 27.—George K. Asymmetry of L5 arch, cleft in S1 arch, wide defect in lower sacral arches.

The first is that of a spina bifida occulta involving the first and the lower sacral arches (Fig. 27), probably not associated with the symptoms:

George K., aged forty-six years, was in the neurological service of Mount Sinai Hospital January 17 to January 31, 1917, with acroparesthesia of the hands and feet.

Following a fall on his back he had suffered for ten years with radiating pains in the lower extremities, varying with the weather. For ten months he had continuous burning pain in the tongue, hands and feet. His hands and feet swelled when dependent.

He had paresthesia ("pins and needles") in the feet. For five years he had impotence and prostatorrhea.

There was decided hyperesthesia of the legs, anteriorly and the buttocks. Tendon reflexes of all four extremities much exaggerated. Lumbar puncture—normal cerebrospinal fluid. Slight polycythemia. Palpation of the sacrum suggests an absence of the lower spinous processes. Other data negative. Roentgenogram shows a spina bifida occulta, illustrated in Fig. 27.

The following case is interesting in the differential diagnosis of the ulceration and gangrene of the toes, the peculiar anomaly of the lower vertebral arches, and in the operative finding:

Isidore B., Russian, male, aged twenty-eight years, was admitted to Dr. Beer's service at Mount Sinai Hospital in August, 1916, with moist gangrene of the fourth and fifth toes of the right foot and mild cellulitis of the anterior portion of the foot. For two years there had been progressive infection, ulceration, and gangrene of the third, second, and first toes of this foot, with failure of the stumps to heal and extension of the process to the plantar surface. The involvement of the fourth and fifth toes had begun three months before admission. During all this time he suffered *severe pain*, which is characteristic of thrombo-angiitis and not of the trophic gangrene of spina bifida.

On the left side the dorsalis pedis and posterior tibial arteries pulsated well, but on the affected side no pulse could be felt in either vessel. The superficial and deep reflexes of all extremities were exaggerated, especially in the lower extremities. No sensory disturbances were elicited. Wassermann reaction negative.

Dr. Abrahamson noted a superficial cleft in the tips of the spinous processes of the eleventh and twelfth dorsal and first and second lumbar vertebræ. A roentgenogram revealed the peculiar deformity of the fifth lumbar and first sacral arches shown in Fig. 28. Although there is no actual cleft in any arch, this type of lamina malformation is seen, as I have shown, in other cases with bifidity; it produces a distinct hiatus vertically, and I think the abnormality may be properly grouped with spina bifida occulta.

Although the patient's disease was fairly diagnosticable as a thrombo-angiitis obliterans, it seemed worth while to give him the benefit of a doubt suggested by the vertebral deformity and to explore it for a possible cauda equina lesion. Dr. Buerger kindly transferred the patient to my care for that purpose.

Operation (Brickner) September 8, 1916. The fifth lumbar and first sacral arches were exposed and found abnormally separated. They were removed, the dura was incised, and the cauda equina was exposed in its bed of fat. It presented no gross lesion. The dura appeared in all respects normal. Spinal fluid escaped under great pressure.

The wound healed *per primam*, and after the operation there was a remission in the subjective symptoms. The second toe was

amputated September 19. There has been a progression of the disease, and the foot will probably require amputation.

The amputated toe showed "chronic suppurative inflammation, granulation tissue, and endarteritis. A diagnosis of primary arterial lesion cannot be determined from the specimen" (Buerger).

FIG. 28.—Isidore B. Malformation of L5 and S1 arches producing a longitudinal hiatus.

Taken all together, the results of operation for spina bifida occulta are not brilliant, and probably for the reason that the degenerative and neoplastic processes are scarcely remediable. Those cases in which there is a hernia of the spinal roots probably offer the best chance for a good result; for the reduction of this hernia into the bony canal, by relieving the contained nerve roots of traction and pressure, may greatly improve the existing symptoms or prevent the development of some others. The third case, Sam S., the young man with the trophic ulcers of the buttocks, represents, I think, a highly satisfactory result from such an operation. Even when there is no meningocele much may be found possible at operation—for example, separation of adherent nerve roots from the membrana reuniens, division of a constricting band, removal

of an endostosis, or *perhaps* even the removal of a teratoma of the cauda equina.

I believe that the following are legitimate indications for operation:

1. In infants and children, spina bifida occulta with congenital lipoma or hypertrichosis, even though without any symptoms—to reduce the spinal hernia into the canal or to meet any other indication that is found, in the hope of *obviating* the development of symptoms during adolescence. The case of the infant that I have reported is illustrative of this indication.

2. In adults, spina bifida occulta with sufficiently serious and especially with progressive symptoms, and this I believe should apply whether or not the spina bifida occulta is marked by external signs (lipoma, hypertrichosis). I am not unmindful of the inherent risks of such an operation, but surely progressive gangrene of the lower extremities and incontinence of the sphincters are indications sufficient to justify the taking of such risks. In the literature I have found records of 12 operations. I am adding now 5 to these—*all of the 17 without mortality*, and some with decided benefit. In all 5 operations for spina bifida occulta within my personal experience there was no untoward result, and the wounds in all cases healed *per primam* and without any spinal infection.

In making this presentation of a condition that has for so many years interested me, it has been my purpose to call attention to the four groups into which it seems to me we may clinically divide spina bifida occulta, and to suggest the advisability of a roentgenographic examination for such a condition in all cases of sphincteric, sensory, trophic, or motor disturbances that might thus be explained, even though there is no external manifestation to suggest its presence.

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